A midline, posterior cervical mass was found during a physical examination of a full-term female infant (Figure 1). The mass was tubular with a wide sessile base and measured $3 \times 3 \times 4$ cm. The base and most of the cylindrical wall were covered by a full thickness of skin, while the dome was covered by a violaceous, tough membrane. No leakage of fluid from the mass was observed and the infant's neurological examination results appeared to be normal.

Figure 2 is a horizontal cut at the C5 vertebral level of the spine using computed tomography. Figure 3 is a sagittal view of the cervical spine using magnetic resonance imaging.

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Denouement and Discussion

Cervical Myelomeningocele

Figure 1. Skin-covered mass arises from the posterior cervical area.

Figure 2. Computed tomographic scan of the spinal cord at the C5 vertebral level shows the extradural component of the myelomeningocele (3). The spinal cord (1) and spinal canal (2) are identified.

Figure 3. Magnetic resonance imaging scan of the cervical spine with the extradural (black arrow) and intradural (white arrow) components of the myelomeningocele identified.

Cervical myelomeningoceles are uncommon dysraphic lesions and comprise a small proportion of neural tube anomalies. They account for 4% to 8% of all myelomeningoceles. 1 The anomaly is usually obvious at birth, presenting as a protruding mass, most often midline. Occult dysraphic lesions may be associated with the Klippel-Feil syndrome, dermal sinuses, or overlying cutaneous changes such as hemangiomas or port-wine stains.

Cervical myelomeningoceles are always covered by a full thickness of skin at the base and a thickened membrane at the dome. 2 This appearance is much different than myelomeningoceles presenting in the thoracic or lumbar areas that tend to be covered by thin, easily ruptured covers. Leakage of cerebrospinal fluid is rare in cervical lesions, as opposed to those that arise more caudal.

TYPES OF LESIONS

Based on anatomic findings, cervical dysraphic lesions are divided into 2 types. The first, known as a myelocystocele, is characterized by a cyst that maintains continuity with the distended central cervical canal and herniates posteriorly into a meningocele. The wall of the myelocystocele contains neurons, ganglia, and glial tissue, while the meningocele that surrounds this cyst is composed of arachnoid and fibrous tissue and is covered by skin. 1 The second type is known as a meningocele, a single cystic sac. A band of tissue that tents the posterior aspect of the cervical cord extends into the wall of this sac. 1

NEUROLOGIC PROGNOSIS

Although most infants with a cervical myelomeningocele may not have detectable neurologic defects at birth or in the early years of life, they almost invariably deteriorate neurologically over time. 2 Two dysraphic malformations of the underlying spinal cord are associated with cervical myelomeningoceles. The more common is the fibroneuromuscular stalk extending from the cervical cord that fans out into the lining of the sac, and the less common one is a split cord malformation in which 2 hemicords are separated by a midline fibrous septum. 3 The 2 malformations associated with the cervical myelomeningocele create tethers that lead to neurologic dysfunction with the growth of the infant or child, sometimes not presenting until adulthood. 3

Whereas Chiari II malformations are almost universally associated with open neural tube defects in the lumbar region, this malformation has been reported in 44% to 62% of the patients with cervical dysraphic lesions. 2 Hydrocephalus, which is usually associated with Chiari II malformations in lumbar lesions, does not consistently correlate with this malformation in the cervical area. 2

RADIOLOGIC INVESTIGATION

Because the long-term neurologic prognosis of infants with cervical myelomeningoceles is dependent on discovering and relieving the tethering effects on the spinal cord that are associated with them, careful neurologic investigation is mandatory. Magnetic resonance imaging provides a means of surveying the neuraxis for associated lesions, but computed tomographic myelography is more sensitive in identifying median fibrous septa and fibroneuromuscular stalks. 2

SURGICAL INTERVENTION

Rather than simply excising the protruding cervical lesion and ligating the dural fistula associated with these lesions, laminectomy, intradural exploration, and resection of all tethering bands and septa should be the definitive initial treatment. 2

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